COMPUTED TOMOGRAPHY IN RETINOBLASTOMA : CT AND HISTOPATHOLOGICAL CORRELATION

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ABSTRACT

Computed Tomography (CT) scan is a valuable adjunct in the differential diagnosis and management of retinoblastoma. Retrospective study of the known retinoblastoma patients with CT scans performed prior to therapy between January 1994 - December 1996 are studied.

18 of 24 Retinoblastoma patients study with CT scan had intraocular calcification demonstrated in at least one eye. 100% of tumors in this study showed evidence of calcification that help much in the diagnosis of retinoblastoma especially in patients under three years old who account for 87.5% of patients. In addition, orbital and brain CT scan performed in all patients are useful in the diagnosis of retrobulbar extension, intracranial seedling, including brain metastasis. Intraparotid gland metastasis in one case, which has no report in any literature, is also found in this study.

INTRODUCTION

Retinoblastoma is the most common intraocular malignant tumor in children. The tumor usually arises from the inner retinal layers and extends as a fleshy nodular mass in the vitreous cavity. The most common route of extraocular extension is along optic nerve, and extension into retrobulbar space or into subarachnoid space. Detection when the disease is confined to the globe is of utmost importance for local control.[1] It is sometimes difficult to diagnose due to a variety of simulating lesions such as coat's disease, primary hyperplastic primary vitreous (PHPV), toxocariasis. Retinal detachment associated with retrolental fibrosis, retinopathy of prematurity must be excluded. To serve this purpose, knowing that the DNA released from necrotic cells in retinoblastoma having a propensity to form a DNA - Calcium complex is usually detected in approximately 95% of histologically examined retinoblastoma.[2] Among the diagnostic tools, CT scan is one of the diagnostic tools that is sensitive in the detection of calcification. Therefore, role of CT in the detection of intraocular calcification and diagnosis is obvious. Not only for detecting the intraocular calcification but also for delineating the retroorbital extent of the tumor and determining if the optic nerve is involved or if there is an intracranial extension when the CT scan is indicated.

MATERIALS AND METHODS

- Twenty four patients (13 males and 11 females) with histopathologically proven to be retinoblastoma with thin section CT scan performed prior to therapy between 1994 and 1996

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were studied. In these cases, 2 mm. contiguous axial scans are obtained through orbits, with thicker sections through the rest of the head.

- Eighteen of the 24 patients had unilateral involvement and 6 had bilateral tumors. The patients' ages ranged from 2 months to 5 years, with a mean of 2.1 years. All patients with unilateral retinoblastoma had their affected eyes enucleated as well as patients with bilateral retinoblastoma had both eyes enucleated. All enucleated eyes were examined histologically.

- By correlative study of the histopathological findings and the image findings from CT scan, we tried to determine the accuracy of CT scan in the detection of intraocular calcification, uveal-scleral tissue involvement, optic nerve involvement, and extraocular extension. In addition, benefits of CT scan in the detection of intracranial metastasis is also documented.

RESULTS

Among 24 patients, 6 patients enucleated from other hospital with no available preoperative CT scan were excluded from this study. Therefore, 18 patients (21 eyes of retinoblastoma) had been studied.

All 18 patients (21 eyes) showed intraocular calcification from CT scan. The patterns of calcification are varied as single and small (Fig. 1A), single and large (Fig. 1B) or multiple and punctate (Fig. 1C). (Table 1)

The choroidal invasion were studied by using the criteria of irregularity, thickness of the uveal - scleral tissue more than 2 mm. (Fig. 2). However, the accuracy in the detection is quite low about 52% accuracy. (Table 2)

Table 1 Intraocular calcification

		Pathological findings	
		positive	negative
CT findings	positive	21	0
	negative	0	0

Table 2 Uveal - scleral involvement

		Pathological findings	
		positive	negative
CT findings	positive	9	8
	negative	2	2

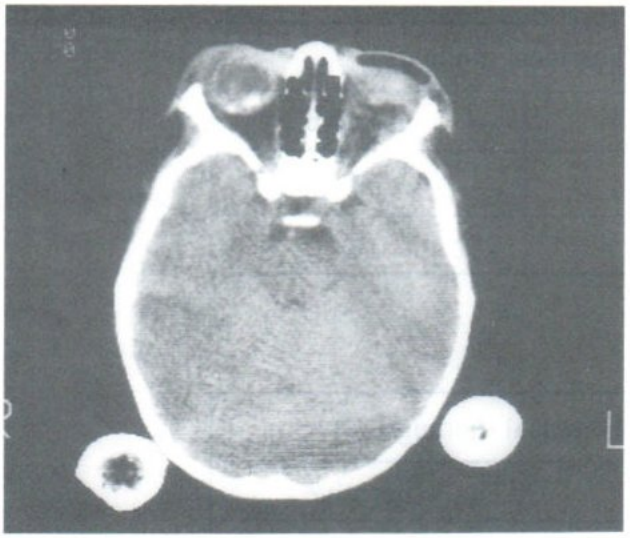
Table 3 Retrobulbar involvement

		Pathological findings	
		positive	negative
CT findings	positive	2	0
	negative	0	19

Table 4 Optic nerve involvement

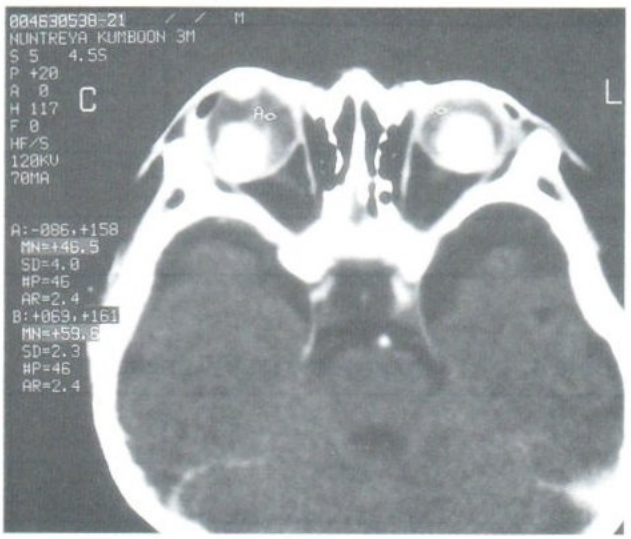
		Pathological findings	
		positive	negative
CT findings	positive	7	0
	negative	4	10

In contrary to the choroidal invasion, the detection of retrobulbar involvement is easily detectable from the evidence of mass that replaced normal retrobulbar fat space (Fig. 3). In our study the accuracy is 100% (Table 3). The optic nerve involvement is demonstrated by CT scan by observing of the enlargement of the optic nerve and/or perioptic enhancement (Fig. 4). Those findings are 83% accuracy (Table 4). Four patients had microscopical evidence of tumor spread past through the lamina cribosa of optic nerve and not reaching the transsection line. This was not detected by CT scan. There were five patients (22%) who had intracranial spreading of tumor. They presented in a normal pattern of CSF spreading with or without hydrocephalus in four cases and an unusual pattern as intraparenchymal mass in one patient (Fig 5, 6). In addition, two patients of those intracranial spread had MRI spines work up for the evaluation of intraspinal metastases. Both of them are positive for intraspinal metastases by the MRI findings which are clearly demonstrated in sagittal post gadolinium contrast study (Fig 7, 8). Moreover, there was a patient who had bilateral retinoblastoma with a subsequent right parotid gland metastasis (Fig. 9). No patient had an evidence of pulmonary metastasis. Only one patient had bony metastasis that involved long bone of lower extremity (Fig. 10).



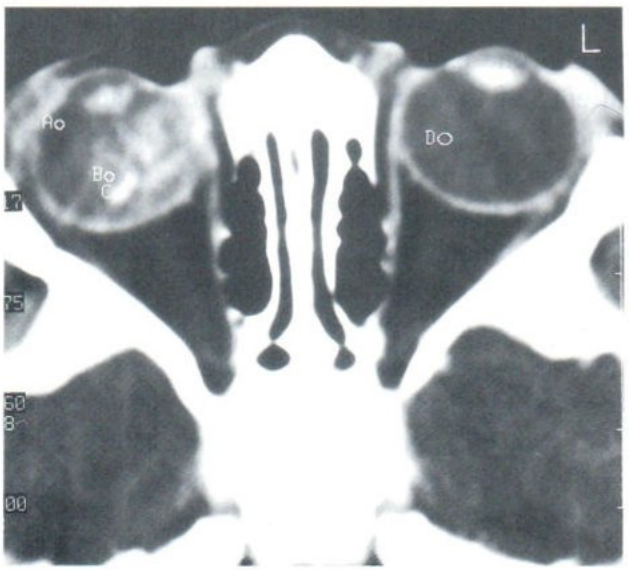
1A

Fig. 1A. This is a case of bilateral retinoblastoma scanning post enucleation of the left eye. The tumor calcification in the right eye is small in size.



1B

Fig. 1B. This is a bilateral retinoblastoma patient which has a large single tumor calcification pattern.



1C

Fig. 1C Multiple punctate intraocular tumor calcification is a pattern of calcification in retinoblastoma.

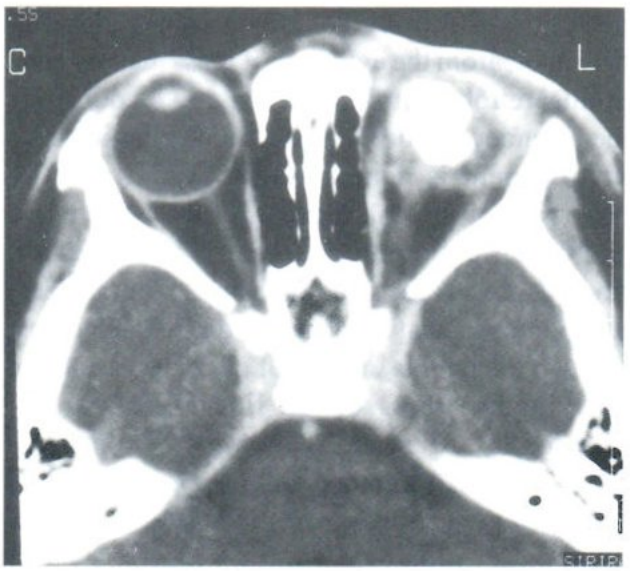


Fig. 2 This is a unilateral retinoblastoma with uveal-scleral tissue invasion. CT depicted as irregular, nonuniform thickening of ocular wall of left eye.

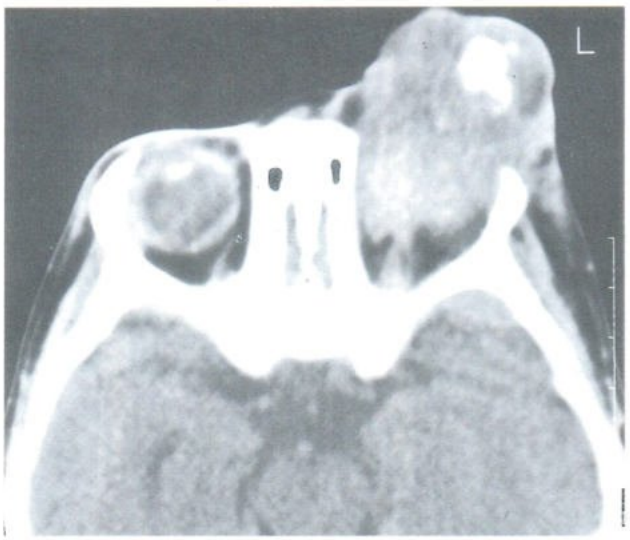


Fig. 3 Unilateral retinoblastoma in left eye with extraocular extension especially retro-bulbar spread

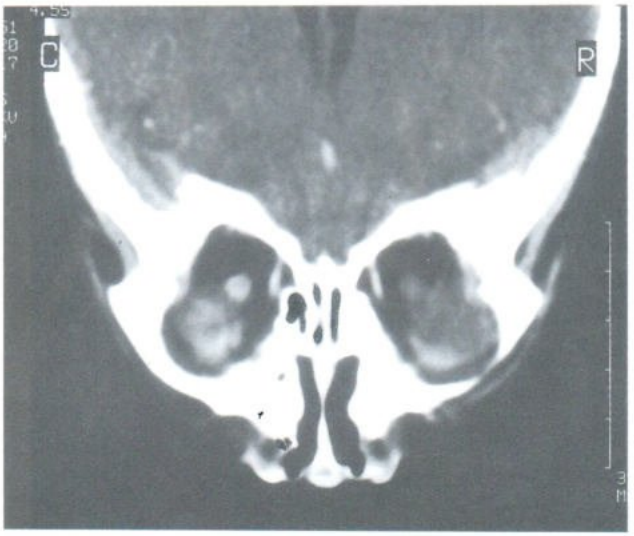


Fig. 4 Contrast enhanced CT brain study in coronal plane reveals asymmetrical enlargement, enhancement of the left optic nerve in a case of retinoblastoma left eye which is suggestive of tumor extension along the optic nerve.

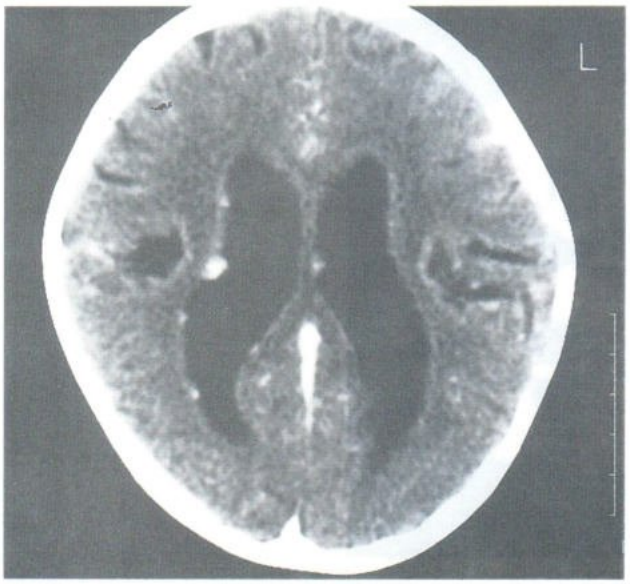


Fig. 5 Contrast enhanced CT brain scan shows multiple ependymal nodules seedling with increase enhancement along the venticular wall and leptomeninges linining of the brain (an example of CSF seedling).

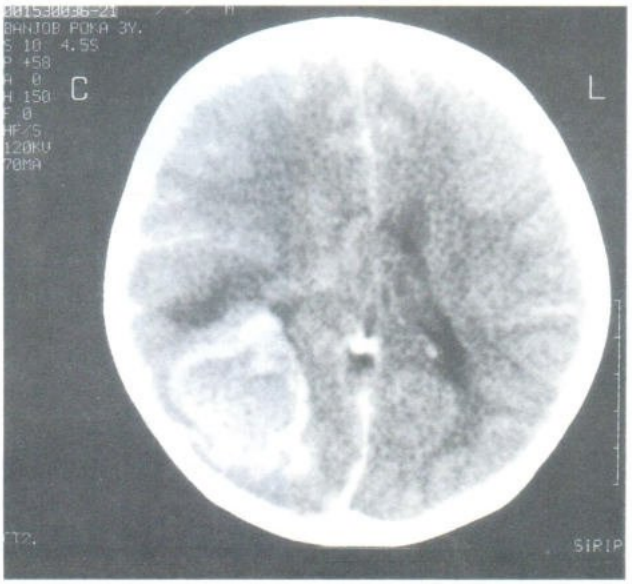


Fig. 6 Contrast enhanced CT brain scan in a known bilateral retinoblastoma, reveals intrapa-renchyma brain metastasis in the right temporo-occipital lobe.

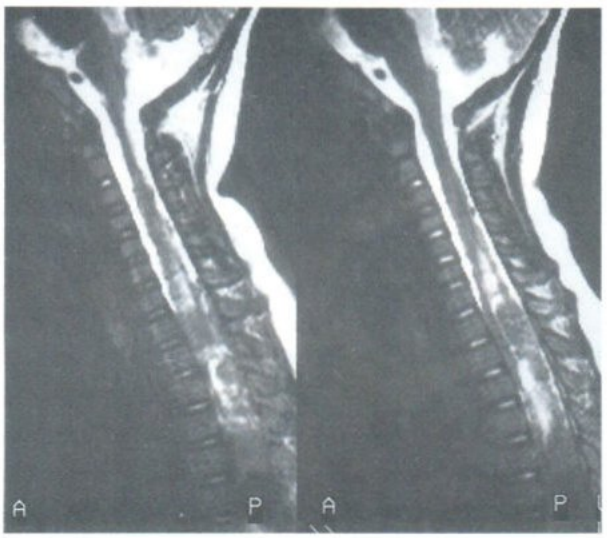


Fig. 7 MRI of cervical spines in bilateral retinoblastoma shows intramedullary metastais of cervical cord.

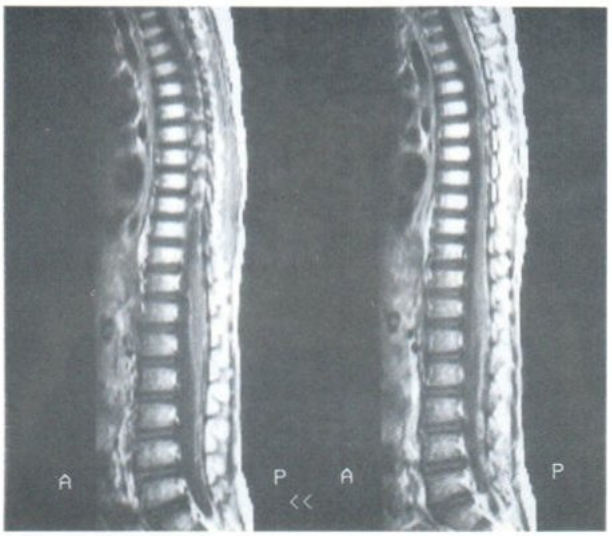


Fig. 8 Gadolinium enhanced MRI of lumbosacral spines showed an abnormal increase enhancement of the dural lining of spinal cord suggestive of tumor seedling along CSF space.

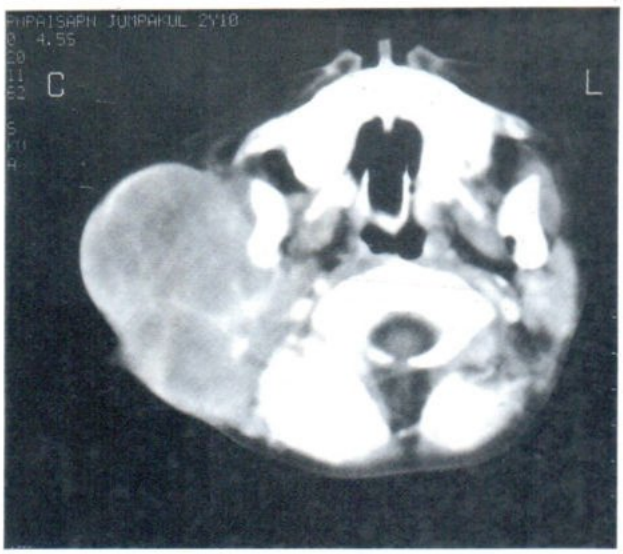


Fig. 9 Contrast enhanced CT study reveals asymmetrical enlargement of the right parotid gland, biopsy proven to be retinoblastoma metastasis.

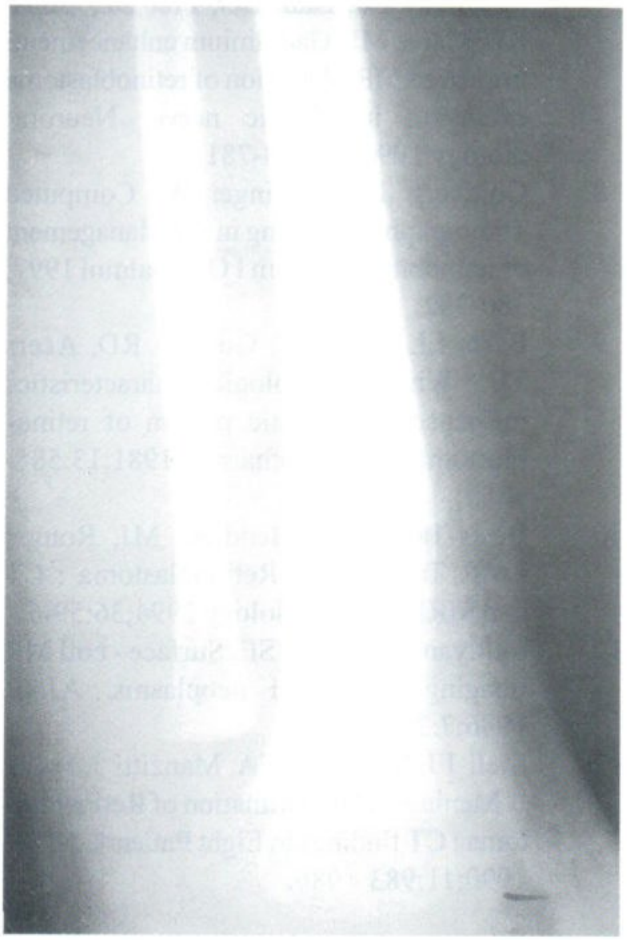


Fig. 10 Plain radiography of the leg in retinoblastoma patient reveals metastatic bone destruction involving anterior aspect of proximal tibia with overlying soft tissue swelling.

DISCUSSION

Computed Tomography can detect calcification within retinoblastomas with a high degree of accuracy. The presence o absence of calcification on the CT examination of the eyes of a patient less than three years of age is an important finding in the diagnosis and the differentiation from other simulating lesions. None of the simulating lesions including optic nerve head drusen, retinal astrocytoma, choroidal hemangioma with previous hemorrhage, choroidal osteoma, toxocariasis, persistent hyperplastic primary vitreous, Coat's disease, retinal dysplasia or trauma tend to contain calcification in the age group (0-3 years) in which retinoblastoma is usually diagnosed.[3-6] Our study confirms the usefulness of the computed tomography in the detection of intraocular calcification with high accuracy as well as the retrobullar extension. The accuracy of optic nerve involvement is quite satisfactory. However, to improve accuracy, MRI (Magnetic Resonance Imaging) with Gadolinium enhancement may be helpful.[7] In term of choroidal invasion, it is very difficult for CT scan to evaluate whether choroidal invasion is present in case of no extraocular extension. It is a fact that the posterior ocular wall composed of retinal, choroidal and scleral layer from inner to outer wall respectively. These structures are not more than 2 mm. in thickness. We attempt to evaluate the tumor invasion by a criteria of asymmetry and/or focal thickening of ocular wall. However, the accuracy is not satisfactory. This could be a limitation of the CT. The other advantages from CT study in the evaluation of the intracranial metastasis are reported.[8,9,12] Our study confirms previous studies,[8,9] in this issue. There are four cases of intracranial metastases, some of these show intraparenchymal metastases. Two of them have intraspinal spreading. In addition, our study shows a patient with bilateral retinoblastoma after enucleation of both eyes coming with right parotid gland metastasis. This was proven by histological section and was an example of lymphatic metastasis. In this decade, eventhough MRI (Magnetic Resonance Imaging) also plays role in the diagnosis of intraocular lesions. We agree with other authors[10,11] suggesting that MRI was not as specific as CT for the diagnosis of retinoblastoma. The superiority of the MRI to CT scan is the ability in the differentiation of Coat's disease from retinoblastoma and subretinal fluid from tumor.[10] We believe that CT is still the method of choice in the diagnosis of retinoblastoma, but when equivocal arises, MRI should be performed for a better differentiation from other

lesions such as Coat's disease[10] and for the improvement of the detection of optic nerve extension.[7]

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